

## Absent Sixth Aortic Arch: A Form of Pulmonary Atresia\*

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Pulmonary atresia is characterized by developmental abnormalities of the pulmonary valve or pulmonary arterial tree, with complete obstruction to blood flow by the normal route. We recognize three main types, based on embryology, anatomy, and suitability for surgical treatment. Pulmonary atresia as an element of a more complex lesion such as transposition of the great vessels or tricuspid atresia is classified under the primary abnormality.

(1) *Pulmonary Valve Fusion with Intact Interventricular Septum*: embryologically this is the extreme form of simple pulmonary valve stenosis, with complete fusion of the pulmonary valve cusps. An interatrial communication is present, usually a patent foramen ovale, and blood reaches the pulmonary arterial tree distal to the obstructed pulmonary valve by way of a patent ductus arteriosus.

(2) *Extreme Fallot's Tetralogy with Pulmonary Atresia*: basically the anatomy is that of the tetralogy of Fallot, with a large ventricular septal defect and dextroposed aorta, but the pulmonary valve and sometimes the main pulmonary artery as well are poorly developed and totally obstructed. Right and left branches of the pulmonary artery are present and communicate with each other across the midline. Blood reaches these vessels via a patent ductus arteriosus or bronchial collateral vessels or both. Taussig (1947) originally confined this term to cases with a functioning patent ductus arteriosus, but we extend it to those with bronchial collateral vessels as an additional or alternative source of blood supply to the branches of the pulmonary artery.

(3) *Absent Sixth Aortic Arch*: we have applied this name to cases in which the entire pulmonary arterial tree fails to develop or is obliterated. In particular the derivatives of the sixth aortic arch of foetal life, the right and left branches of the pulmonary artery, and the ductus arteriosus, are absent or completely atretic. A large ventricular septal defect is present, and the aorta is dextroposed. Blood reaches the lungs by way of one or several large bronchial vessels arising from the descending aorta distal to the origin of the subclavian artery. The present communication is concerned with this group.

### CLINICAL FEATURES

Fifteen cases were available for analysis, with angiographic confirmation in 13 and post-mortem proof in 4. One patient had two thoracotomies carried out in units other than our own.

The age at presentation varied from 1 month to 9 years, 7 being under 6 months and the remainder evenly distributed over the remaining age-groups. Twelve patients were male and three were female, showing a male dominance in contrast with the equal sex incidence in other forms of pulmonary atresia.

The general nutrition was normal in 10 and poor in 5. Disability paralleled the degree of cyanosis, which was slight in 8, moderate in 3, and severe in 4. Haemoglobin varied from 11.9 g./100 ml. to 22.0 g./100 ml., being less than 15 g./100 ml. in 5 and more than 18 g./100 ml. in 3, with intermediate values in the remainder.

The peripheral arterial pulses were normal in 12 and increased in amplitude in 3 patients. In 2 of the latter, episodes of congestive cardiac failure occurred. Increased right ventricular activity on palpation was noted in most patients, but was slight in the majority. Continuous murmurs were present in all, being loud in 10, moderate in intensity in 3, and soft in the remaining 2 patients. The murmurs were widespread over the right side of the chest and the

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back in most patients, and occasionally were heard over the left side of the chest anteriorly as well.

The electrocardiogram showed P pulmonale in 3 instances and normal P wave patterns in the remainder. Right axis deviation was present in 10: slight in 5, and moderate in 5. The QRS axis was within the normal range in the remainder. In the chest leads, slight to moderate right ventricular preponderance was present in 11, 3 were within normal limits, and there was some evidence of hypertrophy of both ventricles in the remaining patient.

#### RADIOLOGICAL EXAMINATION

Plain x-ray films of the chest showed heart size within normal limits in 2 patients, slightly increased in 12, and considerably increased in the remaining patient. The ascending aorta and aortic arch were moderately increased in size in 8 patients, considerably dilated in 5, and within normal limits in 2. The aortic arch was right-sided in 9 patients and left-sided in 6. The lung fields in all patients showed the characteristic appearance of bronchial arterial supply to the lungs as described by Campbell and Gardner (1950) and right and left branches of the pulmonary artery could not be recognized in any. The over-all vascularity of the lung fields was judged to be increased in 3, diminished in 3, and within normal limits in the remaining 9 patients. We seldom carry out barium swallow in the oblique position in our unit, but this method will often demonstrate indentations of the barium-filled oesophagus by large abnormal bronchial vessels, as mentioned by Powell and Hiller (1957).

Cardiac catheterization and selective angiocardiology were carried out in 10 patients, peripheral venous angiograms in 2, and retrograde aortography in 1 patient. Pressures in the right ventricle were at systemic levels in all instances, with a rise in oxygen saturation of varying degree in the same chamber. Peripheral arterial oxygen saturation, where estimated, was moderately reduced.

Selective angiocardiology with injection of contrast material into the right ventricle is the procedure of choice in establishing the diagnosis and excluding other possibilities, and we use simultaneous anteroposterior and lateral views with an Elema-Schonander rapid film changer and cut film. If the blood supply to the lungs is not well demonstrated by this method, retrograde aortography with injection of contrast material just below the origin of the subclavian artery will often give a more precise picture.

Eight angiocardigrams with satisfactory detail were available for review. A large ventricular septal

defect was demonstrated in all. The aorta arose mainly from the left ventricle in 2 patients and astride the 2 ventricles in 6. The aortic arch was right-sided in 5 and left-sided in 3 patients. No elements of the pulmonary arterial system nor a ductus arteriosus could be demonstrated in any. Abnormal bronchial vessels of appreciable size arising from the descending aorta distal to the origin of the subclavian artery were present in all (Fig. 1 and 2). These vessels were of large size in 3 patients, of moderate size in 2, and of small size in 3 patients. Two major arteries could be seen entering the right lung in all 8 patients, and on the left side there were 2 major vessels in 5 and 1 in the remaining 3 patients.

#### NECROPSY MATERIAL

Five patients are known to have died, and the results of necropsy were available in four.

*Case 1*, male, aged 4 months. During life this child was slightly cyanosed, with arterial pulses collapsing in type. Continuous murmurs were widely heard over the praecordium and back. Hb 14.0 g./100ml., an electrocardiogram showed slight right ventricular preponderance, and x-ray films showed cardiac enlargement and increased vascular markings in the lungs. At necropsy the heart was enlarged, with right ventricular hypertrophy and dilatation of both atria. The tricuspid, mitral, and aortic valves were normal. The pulmonary valve, pulmonary artery and its branches, and the ductus arteriosus were absent. There was a high ventricular septal defect and a large overriding aorta, the only outflow from the heart (Fig. 3a). The coronary arteries were normal. The aortic arch was right-sided, giving off a left innominate artery and carotid and subclavian arteries on the right side. From the descending aorta four large bronchial arteries arose, two on each side, to supply the lungs (Fig. 3b).

*Case 2*, male, aged 5 months. In life this patient was undernourished and moderately cyanosed, with widespread continuous murmurs over the right side of the chest and the back. An electrocardiogram was within normal limits and x-ray films showed slight cardiac enlargement and diminished vascular markings in the lungs. At necropsy the heart was enlarged, with some hypertrophy of both ventricles and atria. There was a high ventricular septal defect, just under the common ventricular outlet into the aorta. The mitral and tricuspid valves were normal. The aortic valve had three cusps, but there was one large common coronary artery dividing shortly after its origin into two large branches. The entire pulmonary arterial system was absent. The right aortic arch gave off left innominate and right common carotid and subclavian arteries. Distal to the subclavian artery two bronchial arteries arose, one entering the hilum of each lung.

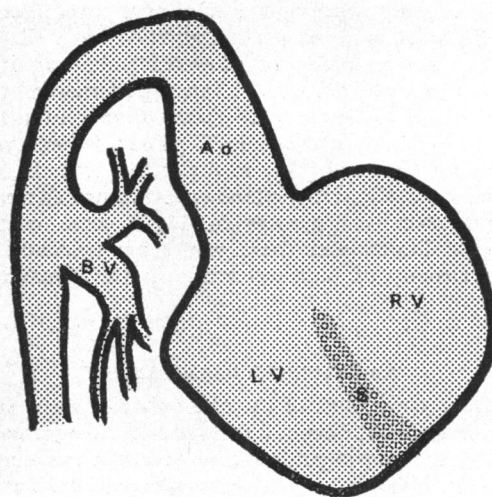
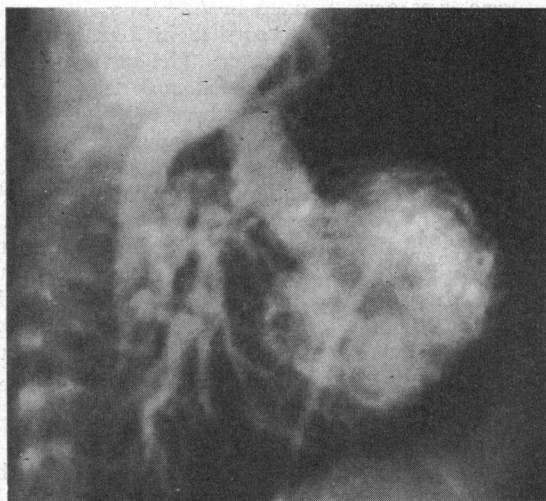


FIG. 1.—Angiocardiogram, lateral projection. Ventricular septal defect demonstrated. Aorta (Ao), arising astride the interventricular septum (S), fills directly from right ventricle. Right ventricular outflow tract and pulmonary artery absent. Several large bronchial vessels (BV) arise from the descending aorta to enter the lungs.

*Case 3*, male, aged 12 months. This child failed to thrive and had repeated respiratory tract infections and congestive cardiac failure. Cyanosis was slight, Hb 11.9 g./100 ml. The arterial pulses were collapsing in character, and continuous murmurs were heard all over the chest anteriorly and at the back. An electrocardiogram showed some evidence of hypertrophy of both ventricles, and on x-ray examination there was considerable

cardiac enlargement and increased vascular markings in the lungs. At necropsy there was general enlargement of the heart, particularly the right-sided chambers. A common truncus arose from both ventricles above a large ventricular septal defect and formed an aorta which arched over the right bronchus (Fig. 4a). Opposite the lung hila, the descending aorta gave rise to a large trunk, from which arose four arteries, three on the right and one

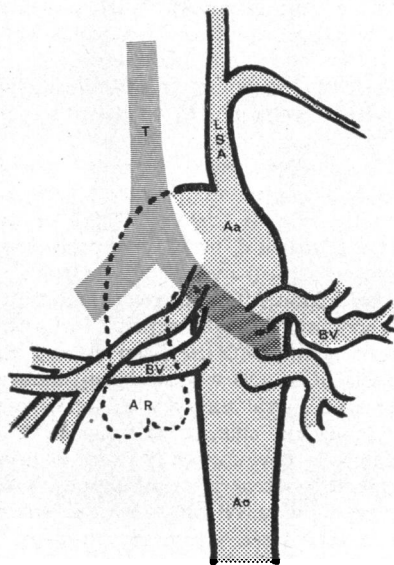
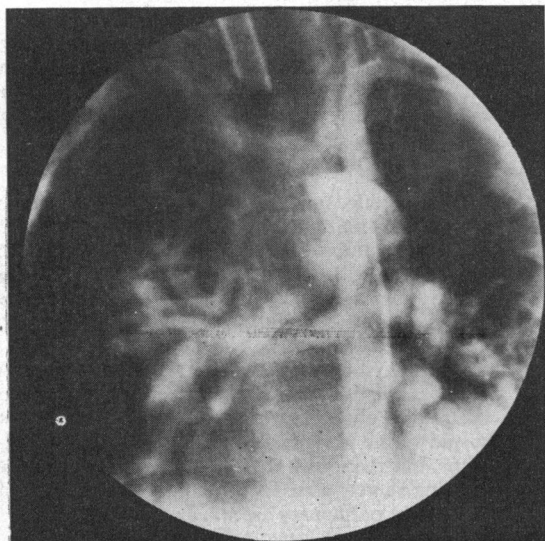


FIG. 2.—Retrograde aortogram. Two very large bronchial vessels (BV) on the left side and two moderate sized vessels on the right side leave the descending aorta (Ao) to supply the lungs. T, trachea; LSA, left subclavian artery; Aa, aortic arch; AR, aortic root.

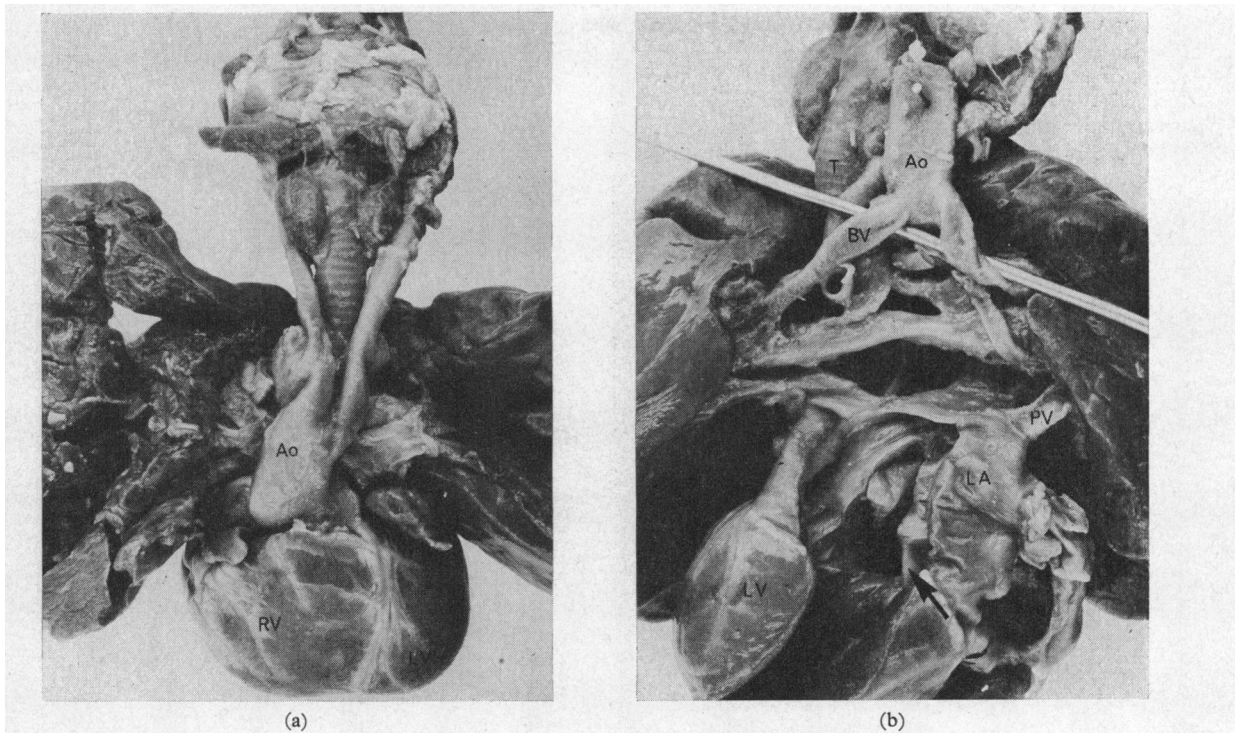


FIG. 3.—Case 1. (a) Anterior view showing a large aortic trunk (Ao) arising from both ventricles, with right-sided arch. Main pulmonary trunk and its branches absent. (b) Posterior view with descending aorta (Ao) pinned upwards to show large bronchial vessels (BV) arising from its ventro-lateral aspect to supply the lungs. T, trachea; PV, pulmonary vein; LA, left atrium; LV, left ventricle. Arrow indicates ventricular septal defect.

on the left, which passed to the lung hila (Fig. 4b). A fibrous strand arose from the base of the heart and passed to the left lung hilum. Shortly above its origin a finer strand arose and passed to the right lung hilum (Fig. 4a). There was no ductus arteriosus.

*Case 4*, female, aged 9 years. This patient was moderately cyanosed, Hb 17.0 g./100ml., with loud continuous murmurs on the right side of the chest and at the back. An electrocardiogram was within normal limits, and on x-ray examination the heart was slightly enlarged, the aorta considerably dilated with left-sided arch, and the vascular markings in the lungs were within normal limits. At necropsy the heart was considerably enlarged and the ascending aorta grossly dilated. The pulmonary artery could not be identified and no ductus was demonstrated. The entire pulmonary blood supply consisted of enlarged bronchial arteries arising from the descending aorta. The left atrium and left ventricle were dilated and thick-walled. The right atrium was small and communicated with the left atrium by a greatly dilated foramen ovale. The tricuspid orifice was stenosed and the right ventricular cavity small. A small ventricular septal defect was present, but no pulmonary orifice.

#### EMBRYOLOGY

Following the description of Huntington (1919), in the developing foetus the pulmonary vascular beds are established independently of the sixth aortic arch. They are connected at an early stage to the ventral branches of the dorsal aortae. Later they send out cranial branches to join the ventral roots of the sixth aortic arch, which form the main branches of the pulmonary artery, and communication with the dorsal aorta is lost. The dorsal root of the sixth arch joins the ventral root just distal to the aortic bulb, and persists as the ductus arteriosus on the left side. The fourth arch persists on the left side as the aortic arch. On the right side the fourth aortic arch persists as the subclavian artery and the dorsal root of the sixth arch disappears. The aortico-pulmonary septum arises from the angle between the fourth and sixth arches, and grows spirally towards the heart, dividing the primitive truncus arteriosus into the aorta and the main pulmonary artery.

The fundamental embryological fault in the condition under discussion is the absence or early involution of the primitive sixth aortic arch. Ventral

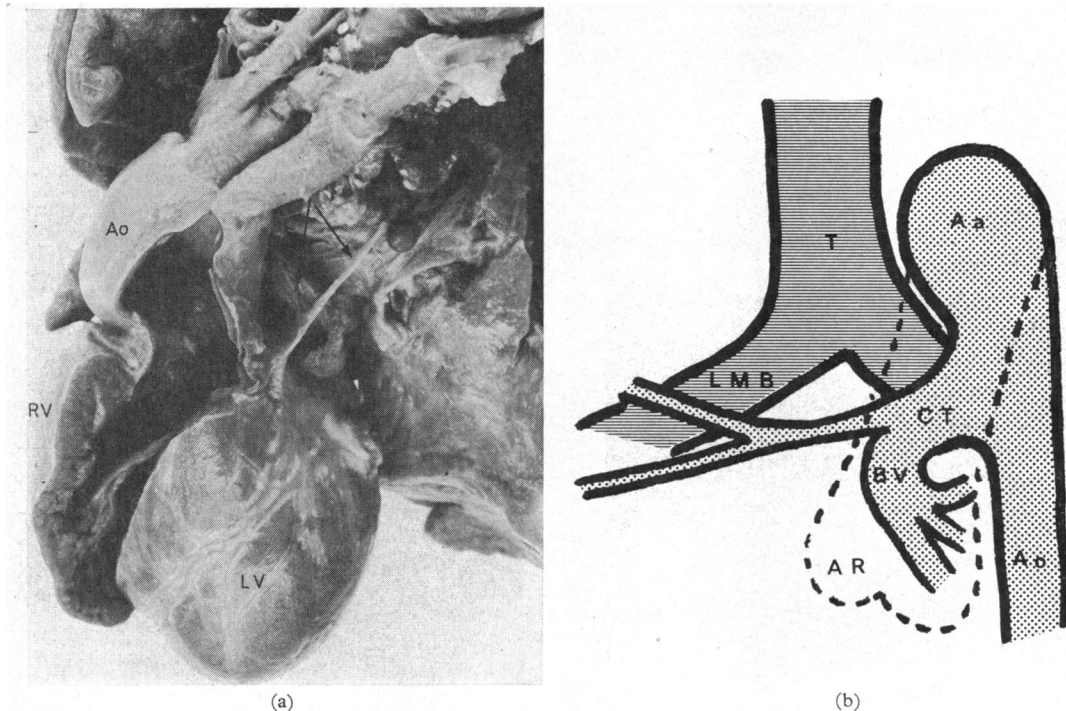


FIG. 4.—Case 3. (a) Anterior view showing a large aortic trunk (Ao) arising astride a large ventricular septal defect. The pulmonary artery is represented by a fibrous strand (arrow) passing upwards to the hilum of the left lung. An even finer fibrous strand (arrow) branches off and passes to the hilum of the right lung. (b) Line drawing of same specimen, posterior view. The descending aorta (Ao) has been displaced to the right to show a large common vascular trunk (CT) leaving its ventral aspect and giving off substantial branches (BV) to each lung. T, trachea; LMB, left main bronchus; Aa, aortic arch, AR, aortic root.

branches of the dorsal aorta persist as the only source of blood supply to the lungs. Complete absence of the sixth arch implies an early embryological fault, before the fourth week of foetal life. The third post-mortem specimen, in which the main pulmonary artery and its branches are represented by fibrous strands, implies arrest and involution at a later stage of development, but again primitive ventral branches of the dorsal aorta persist as the only source of blood supply to the lungs.

#### DIFFERENTIAL DIAGNOSIS

Absent sixth aortic arch must be distinguished from persistent truncus arteriosus on the one hand and from other forms of pulmonary atresia on the other.

As regards persistent truncus arteriosus, we accept the diagnostic criteria suggested by Tandon, Hauck, and Nadas (1963), namely ventricular septal defect, a single vessel forming the outlet for both ventricles and giving rise to the coronary arteries, and origin of both pulmonary arteries from this trunk before any brachiocephalic vessels are given

off. Continuous murmurs are seldom heard in this group, and angiograms will usually demonstrate pulmonary arteries arising either singly or by a common vessel from the persistent truncus.

In pulmonary valve fusion with intact ventricular septum, continuous murmurs, when present, are confined to the usual ductus area. Angiograms show a well-defined infundibulum of the right ventricle ending blindly at valve level, and retrograde filling via a patent ductus of a normal pulmonary arterial system distal to the obstructed pulmonary valve (Grattan-Smith *et al.*, 1966).

The most difficult differentiation during life is from extreme Fallot's tetralogy with pulmonary atresia. Widespread continuous murmurs are heard in these patients, especially when large bronchial collateral vessels are present. The essential point is the angiographic demonstration of right and left pulmonary arteries of normal morphology and often relatively normal size, which communicate with each other across the midline. The pulmonary arteries fill from a patent ductus arteriosus or from large bronchial vessels, or from both.

## DISCUSSION

Examples of absent sixth aortic arch have been reported by previous authors under a confusing variety of names. Taussig (1947) in the first edition of her book spoke of a "pseudo" truncus arteriosus, representing an arrest in the formation of the sixth branchial arch, with blood supply to the lungs by way of collateral circulation, usually bronchial arteries. She also described truncus arteriosus with circulation to the lungs by way of the bronchial arteries, mentioning continuous murmurs limited to one side of the chest, and indentation of the barium-filled oesophagus by large bronchial arteries, yet in the one post-mortem case described, a small pulmonary artery was present which branched to the lungs in the normal fashion, though not communicating with the heart. Mention was also made of truncus arteriosus, with circulation to the lungs by way of the superior bronchial arteries, but again the post-mortem example had a small pulmonary artery branching normally to the lungs, but ending blindly near the base of the heart.

Manhoff and Howe (1949) must be credited with first recognizing this condition as a distinct embryological and anatomical entity, in a paper entitled "Absence of the Pulmonary Artery". They had one post-mortem specimen of their own, and found six similar cases reported before 1949, under different diagnoses. They realized that the condition was due to failure of development of the sixth aortic arch, with persistence of primitive bronchial vessels from the dorsal aorta as the only source of blood supply to the lungs. They were also aware that the condition was compatible with slight cyanosis and disability, and with survival to adult life.

Collett and Edwards (1949), under the heading persistent truncus arteriosus type IV, described cases in which the pulmonary arteries were absent and the arterial circulation to the lungs was by way of bronchial arteries (congenital absence of each sixth arch). In later writings Edwards (Edwards *et al.*, 1965) is inclined to regard this group as embryologically distinct from other forms of persistent truncus arteriosus, and also stresses the major differences from "pseudo-truncus arteriosus", a variant of the tetralogy of Fallot, in which late filling of true pulmonary arteries occurs—extreme Fallot's tetralogy with pulmonary atresia in our terminology.

Campbell and Gardner (1950) mentioned two patients, both investigated angiographically and one surgically, who almost certainly belonged to this group, as persistent truncus arteriosus with bronchial artery supply to the lungs. Their remaining five post-mortem cases were examples of extreme Fallot's tetralogy with pulmonary atresia.

Roche (1953) reported three cyanotic patients with continuous murmurs explored surgically, probably all examples of extreme Fallot's tetralogy with pulmonary atresia. He mentioned a rare primitive type of truncus arteriosus in which the developing pulmonary artery fails to join either ventral or dorsal aorta, with no pulmonary artery branches or ductus arteriosus, and blood supply to the lungs via bronchial vessels. He also recognized that continuous murmurs were heard when blood was conveyed to the lungs by one or a small number of large bronchial arteries.

Powell and Hiller (1957) described 20 patients with cyanosis and widespread continuous murmurs under the title "ducto-pulmonary atresia". This group probably included some with absent sixth aortic arch, but there was no distinction made from extreme Fallot's tetralogy with pulmonary atresia, and right and left branches of the pulmonary artery were thought to be present in a number of their cases. No post-mortem material was available.

Campbell and Deuchar (1961) reported one additional case with typical angiographic and post-mortem findings, under the heading persistent truncus arteriosus Group I (B), with bronchial arteries providing the only blood supply to the lungs, and commented that derivatives of the sixth arch were absent in this group.

The clinical picture in absent sixth aortic arch depends on whether the pulmonary blood supply is inadequate, adequate, or excessive, as judged by the degree of cyanosis and disability, and by the radiological estimate of vascularity of the lung fields. Those with inadequate pulmonary blood supply make poor progress and die in the early months or years of life. Three of our patients were in this group, and Case 2 is an example.

Four of our patients had excessive pulmonary blood supply. It is a curious paradox that this may occur despite absence or complete obstruction of the pulmonary artery. Similar cases in other forms of pulmonary atresia have been reported by Campbell and Deuchar (1961) and by Venables (1964). Cyanosis is slight, the arterial pulses are collapsing in character, and congestive cardiac failure is common. Uncomplicated patent ductus arteriosus may be simulated. Progress is poor and survival limited. Case 3 is an example of this group.

The majority of patients with absent sixth aortic arch, 9 in our series, have an adequate pulmonary blood supply. Cyanosis is slight or moderate, growth and nutrition are normal, and there is remarkably little disability. Our oldest patient remains active and well at 19 years of age, having been under observation for 17 years. This relatively good prognosis has been mentioned by



Manhoff and Howe (1949) and by Powell and Hiller (1957). Perhaps this can be attributed to the developmental origin of the primitive bronchial arteries from tissues destined to cope with systemic pressures throughout a lifetime.

The marked male dominance in absent sixth aortic arch, sufficient to be of clinical diagnostic importance, is of interest and contrasts with the female dominance in persistent patency of the ductus arteriosus. Absence or persistence of the ductus may be sex-linked at the genetic level.

There seems little prospect of surgical treatment for this condition. The bronchial arteries supplying the lungs have origin directly from the aorta and carry high pressures. One of our patients had two unsuccessful thoracotomies carried out in other units. The only operation we have contemplated, but not yet carried out, is multiple banding of bronchial arteries near their origin from the descending aorta in patients with excessive pulmonary blood supply.

#### SUMMARY

Absent sixth aortic arch is a form of pulmonary atresia in which the fundamental lesion is the failure of development or total atresia of the pulmonary arteries and the ductus arteriosus, the derivatives of the sixth aortic arch of foetal life. The sole blood supply to the lungs is by primitive bronchial arteries arising from the descending aorta distal to the origin of the subclavian artery.

Fifteen cases are presented, with angiographic confirmation in 13 and necropsy proof in 4; 12 were male and 3 female, showing male dominance. Cyanosis is always present but may be slight. Continuous murmurs are widely heard over the chest, particularly on the right side and at the back. A right aortic arch is present in more than half the cases. The clinical picture depends on whether the blood supply to the lungs is inadequate, adequate, or excessive, but some patients have little disability and

survive to their second or third decade. An interesting subgroup comprises those with excessive pulmonary blood supply who develop congestive cardiac failure and die in the early months or years of life.

The condition must be distinguished from persistent truncus arteriosus on the one hand, and from other forms of pulmonary atresia on the other. The diagnosis can usually be established during life by selective angiocardiology with right ventricular injection or by retrograde aortography. Surgical treatment is not feasible at the present time.

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